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CASE REPORT

Progressive Encephalomyelitis With Rigidity: A Case Report

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Abstract

Background/Objective: The most prominent clinical features of progressive encephalomyelitis with rigidity (PER) are painful spasms and rigidity accompanied by clinical signs of brainstem and spinal cord involvement. In initial reports, PER had fatal outcome. Later, clinical improvement related to corticosteroid therapy has been described in some cases. The objective of this study was to signify a reputed clinical significance of corticosteroid therapy in PER.

Methods: Case report.

Results: A 50-year-old man developed progressive syndrome of tonic extensor spasms. Magnetic resonance imaging (MRI) showed areas of signal changes in cervical spinal cord and lower brainstem, whereas cerebrospinal fluid analysis indicated subacute encephalomyelitis. His condition dramatically improved on oral corticosteroid therapy. Clinical improvement was accompanied by normalization of MRI findings.

Conclusion: For this patient with PER, corticosteroid therapy was a dramatically effective and life-saving treatment, although initiated rather late in the course of the disease.

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Key Words: Spasticity; Muscle spasms, tonic; Encephalomyelitis, progressive; Corticosteroid therapy; Methylprednisolone; Magnetic resonance imaging; Decerebrate posture

INTRODUCTION

Progressive encephalomyelitis with rigidity (PER) is a rare but well-defined clinical and pathologic syndrome of unknown etiology. The most prominent clinical features of PER are painful spasms and rigidity that are accompanied by clinical signs of brainstem and spinal cord involvement. In initial reports, PER had fatal outcome (1–3); the principal neuropathologic findings were those of subacute encephalomyelitis primarily affecting gray matter of the spinal cord and lower brainstem. In later reports, the efficacy of immunosuppressive therapy was reported in a few cases.

CASE PRESENTATION

A 50-year-old man reported the onset of pain in the left eye and hypoesthesia and spasms on the left side of his face. Computer tomography (CT) brain scan, brain magnetic resonance imaging (MRI), cerebral pan-angiography, and standard hematology and biochemistry tests were normal. Trigeminal symptoms improved on therapy with gabapentin. After a 2-week remission, symptoms progressed further.

One month later, the patient developed pain and loss of sensation in the interscapular region and left shoulder. The following month, he was readmitted because of urinary retention, paraplegia, and marked weakness of the arms. The weakness was flaccid and deep tendon reflexes were diminished. Abdominal reflexes and plantar responses were absent. He had horizontal nystagmus on lateral gaze to the left and allodynia in the territory of the left ophthalmic branch. Sensation below C3 dermatome was impaired, which was more pronounced on the left half of the body. Swallowing and chewing were difficult, and his voice became hoarse. At this stage, there were no involuntary movements.

Laboratory workup was negative, including hematology and biochemistry tests, immunologic tests (including rheumatoid factor, immunoelectrophoresis, complement, circulating immune complexes, anti-nuclear, anti-smooth muscle fiber antibodies, antiphospholipid antibody, anti-neutrophil cytoplasmic autoantibodies, angiotensin-converting enzyme, and visual-evoked potentials and nerve conduction studies). Electromyography revealed no denervation activity. Neoplastic workup was negative.

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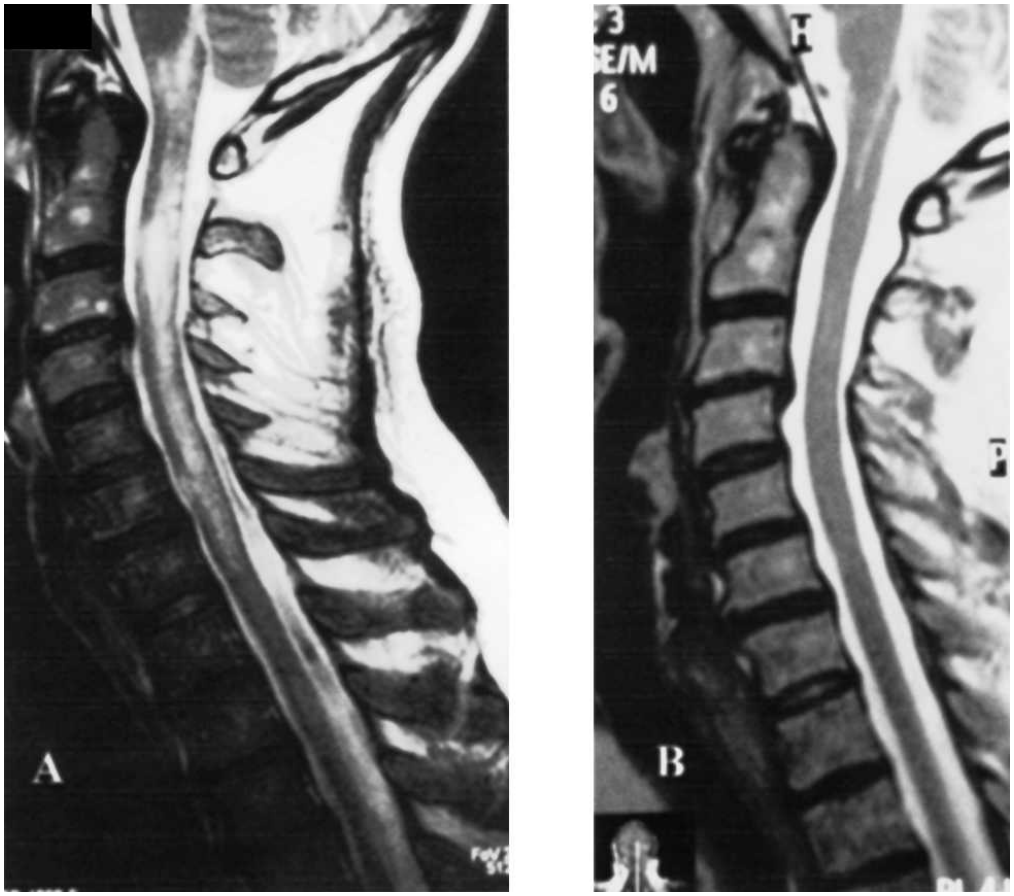

Figure 1. *(A) MRI scan of the cervical cord. Sagittal T2-weighted image showing increased signal intensity of the cervical spinal cord and lower brainstem. (B) Sagittal T2-weighted image 2.5 years later; normal signal intensity.*

MRI of the spinal cord showed an intramedullary lesion expanding the cervical cord, extending from the medulla oblongata to the T1 segment. It was isointense on T1-weighted images and hyperintense on T2-weighted images (Figure I) and showed patchy central and patchy meningeal enhancement with gadolinium. Spinal angiography was normal. Cerebral spinal fluid (CSF) contained 480 white cells/mm^3 (84% lymphocytes) and 1.123 g/L protein. Repeat lumbar puncture (performed 17 days after the initial one) showed CSF with 62 white cells (98% lymphocytes) and 0.65 g/L protein. Electrophoresis findings were unremarkable. IgG index was 0.48. Oligoclonal bands were absent. No viruses were cultured from the CSF, and no intrathecal antibody production against *Borrelia burgdorferi* was detected.

One month later, the patient developed episodes of painful tonic extensor spasms of the upper and lower limbs. The spasms started in the left hand, rapidly spreading to all muscles of the upper and lower extremities. At first, the spasms occurred several times a day and increased in frequency. We were not able to measure aquaporin antibodies, anti-gamma-aminobutyric acid, anti-glutamic acid decarboxylase, and anti-amphiphysin antibodies.

An intravenous course of human immune globulin (0.4 g/kg/24 h for 5 days) was administered with no benefit. Diazepam, baclofen, phenytoin, gabapentin, and morphine were tried in an effort to control the spasms, also with no benefit. He was constantly screaming because of painful tonic spasms, which became almost continuous. The spasms occurred spontaneously or were provoked by various stimuli. They occurred during the day and night, making feeding and sleeping impossible. The general pattern of these painful spasms resembled that of the decerebration syndrome in mesencephalic lesions. The shoulders were drawn forward; the arms were anteflexed at the shoulders, hyperextended at elbows, pronated with hyperflexion of the wrists, and, to a lesser degree, the fingers. The spine was in an exaggerated lordosis. The legs were stiffly extended at hips and knees with extreme plantar flexion of the feet and toes. Figure 2 depicts the pattern of generalized muscle spasms (it is not a photo of the patient, it is a photomontage).

At that time, 5 months after disease onset, therapy with methylprednisolone (64 mg/24 h) was introduced. Favorable response was observed on day 6 of therapy, when the spasms decreased in frequency and severity. After 30 days of treatment, he was able to walk with

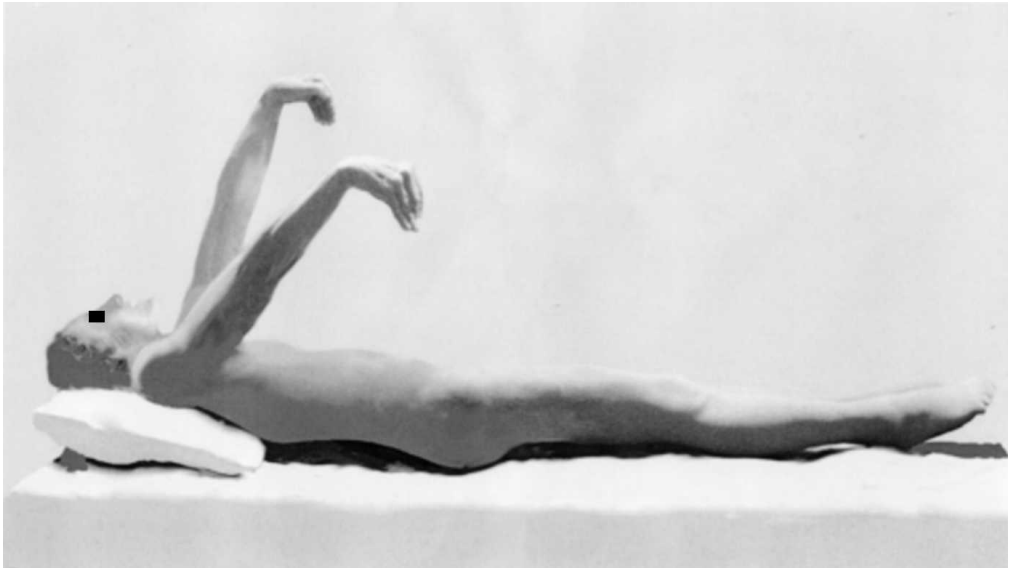

Figure 2. *A depiction of the pattern of generalized muscle spasms (by B. Jušić).*

assistance. When the corticosteroid dosage was gradually reduced to 64 mg/48 h, spasms recurred, and the dosage had to be titrated back to the previous value, resulting in improvement. Corticosteroid therapy was discontinued after more than 2 years of continuous administration. Three years after disease onset, neurologic examination showed only some sensory changes in the left arm, slightly positive Babinski on the left and brisk proprioceptive reflexes of the legs, and no spasms or muscle pain.

DISCUSSION

Based on the histopathologic findings, identification of autoantibodies, and improvement on immunosuppressive therapy in some cases, the concept of autoimmune mechanisms has an important role in the interpretation of PER pathogenesis (4–9). In this case, the clinical, radiological, and laboratory features were very much like those in the case of PER reported by McCombe et al (9). In both cases, limb or trunk rigidity was not recorded, pain and sensory disturbances were present at disease onset, and later weakness developed followed by involuntary movements after several months of progressive neurologic disorder. Because of the simultaneous stiffening of all 4 limbs into the decerebration posture and association with piloerection and hyperventilation in their case, McCombe et al (9) suggested that the brainstem may have been the site of the disturbance. Kasperek and Zebrowski (10) also observed a posture similar to that of decerebration in their patient, as we did in our patient. The mechanism of extensor spasms in these patients has been proven. It could be a release phenomenon because of brainstem dysfunction or the consequence of α-motoneuron disinhibition because of disruption of the spinal inhibitory mechanisms.

Clinical improvement related to corticosteroid therapy has been described elsewhere (6,8,9,11). It is important to note that in our patient corticosteroid therapy had to be continued for a long period of time. After 2 months of treatment, during attempted tapering off of corticosteroid therapy, muscle spasms recurred, and the dosage had to be increased again to the previous level. It was only after more than a year of treatment that the dosage could be significantly reduced without relapse.

Based on experience, Meinck and Thompson (6) suggested that initial treatment with methylprednisolone (500 mg, intravenous, for 5 days) be followed by maintenance oral treatment, being superior to intravenous IgG (30 g/d for 5 days). Intravenous immunoglobulin was not effective in our patient, which is consistent with previous observation. Molina et al (12) reported a person with PER who had an excellent response to intravenous immunoglobulin in association with oral prednisone 80 mg/24 h.

Although MRI is very important in confirming the diagnosis of PER, the lesions may be undetected; the majority of reported cases had normal MRIs (4,5,11,13). Only a few patients have been reported with neuroimaging abnormalities related to their disease (8,9). In our patient and in the case reported by McCombe et al (9), MRI scans showed a cervical cord abnormality extending to the brainstem that was suggestive of myelitis. In both cases, clinical improvement was accompanied by normalization of MRI findings.

CONCLUSIONS

According to the unique clinical features, clinical improvement on corticosteroid treatment, and MRI and CSF findings, this case was classified as encephalomyelitis with PER syndrome. In this patient, corticosteroid therapy was a dramatically effective and life-saving treatment, although initiated rather late in the course of the disease.

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